THE CLASSIC PRESENTATION

of Spinal CSF Leak Symptoms

JILL RAU, MD, PHD, FRCPC, FAHS

THE BOB BOVÉ NEUROSCIENCE INSTITUTE @ HONORHEALTH SCOTTSDALE, AZ

UNIVERSITY OF ARIZONA – PHOENIX, SCHOOL OF MEDICINE

...in case you're just joining us.

DISCLOSURES

- Medical Advisory Board Member of the Spinal CSF Leak Foundation
- Principal Investigator ileak registrySM
 (coming soon)
- The images adorning this presentation were taken from the web and were created by Al and/or very talented people who are not me.







SCHIEVINK CRITERIA

Criterion A

Demonstration of a spinal CSF leak (i.e. presence of extrathecal CSF)

Or, if A not met

Criterion B

Cranial MR imaging changes of intracranial and least one of the following:

- low opening pressure
 (≤60 mm H2O)
- 2. spinal meningeal diverticulum
- 3. improvement of symptoms after EBP

Criterion C

The presence of all of Criterion B features or at least 2 of the features if typical orthostatic headaches are present.

Or, if A and B not met

ICHD-3 7.2 HEADACHE ATTRIBUTED TO LOW CEREBRAL SPINAL FLUID PRESSURE

- Diagnostic criteria:
- 1. Any headache¹ fulfilling criterion C
- 2. Either or both of the following:
 - 1. low CSF pressure (<60 mm CSF)
 - 2. evidence of CSF leakage on imaging²
- 3. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or led to its discovery³
- 4. Not better accounted for by another ICHD-3 diagnosis.

Notes:

- 1. Is usually but not invariably orthostatic (worsens soon after being upright or and/or improves after becoming horizontal)
- Brain imaging showing brain sagging or pachymeningeal enhancement, or spine imaging showing extradural CSF. (spine MRI, or MRI, CT or digital subtraction myelography)
- 3. May depend upon onset in temporal relation to the presumed cause, together with exclusion of other diagnoses.

RELATIVELY UBRUPT ONSET

Very often patients can identify the day that their symptoms onset.

 Less commonly symptoms may wax and wane for a short period of time before becoming more persistent



ORTHOSTATIC SYMPTOMS



orthostatic adjective

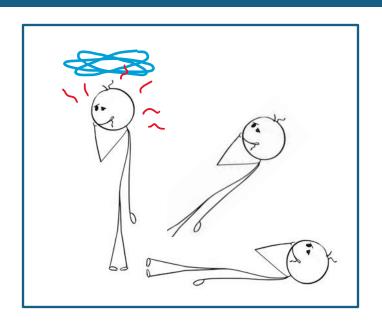
or·tho·stat·ic (ˌor-thə-ˈsta-tik ◄)

: of, relating to, or caused by an upright posture orthostatic hypotension

HEADACHE / ORTHOSTATIC HEADACHE

- 70-100% report headache
 - head pain / head pressure
- Often diffuse or most pronounced at the back of the head



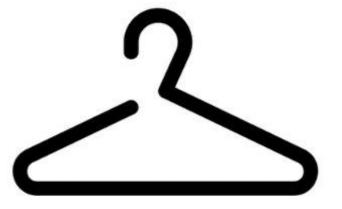


- ~90% orthostatic head pain / headache
- Relieved or significantly improved with lying down, worse with upright
- Second-half-of-the-day headache

NECK PAIN / STIFFNESS



- Neck pain / stiffness is one of the next most common symptoms reported
- Sometimes described in a coat hanger distribution





AUDITORY SYMPTOMS

- Tinnitus
- Aural fullness
- Phonophobia
- Hearing changes (hyperacusis)



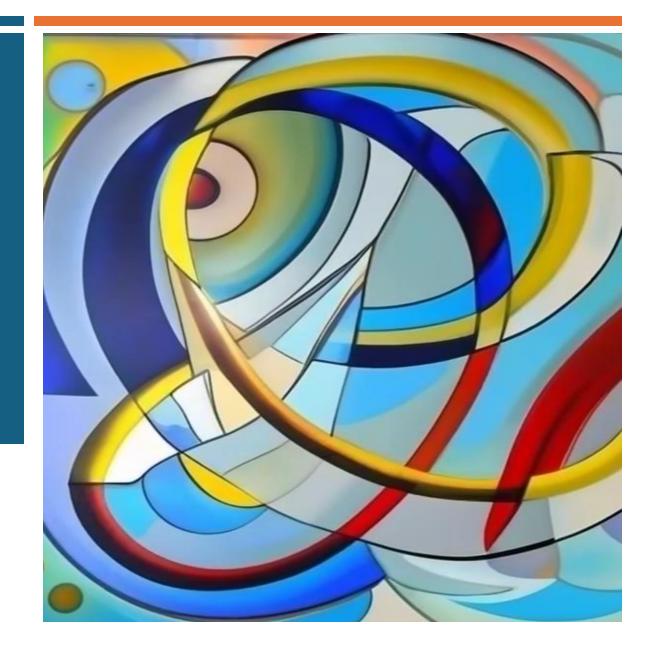
NAUSEA

- +/- VOMITING
- reported 20-60%

DIZZINESS

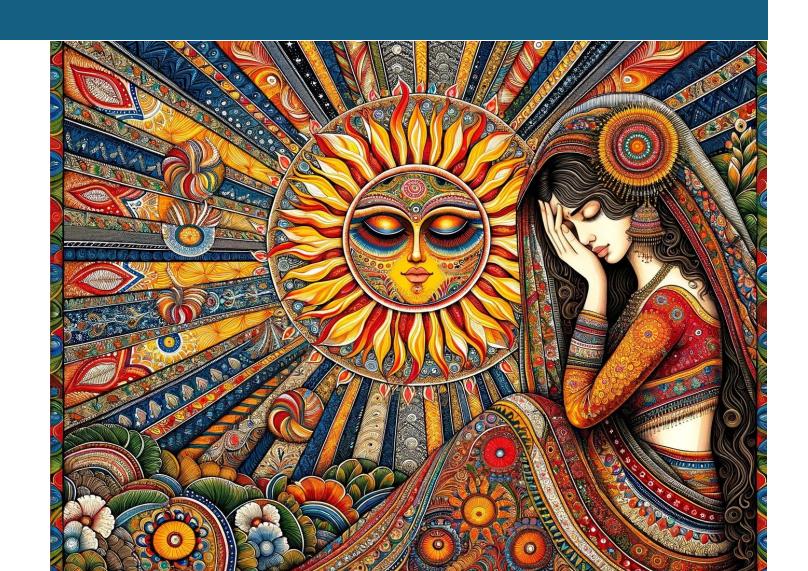
- Reported up to 50%
- Orthostatic, transient
- Lightheadedness
- Dysequilibrium
- Vertigo less common





PHOTOPHOBIA

- Reported in 5 40%
- Less commonly, other visual symptoms
 - Blurry vision
 - Diplopia



BRAIN FOG / NEUROFATIGUE

- Not documented frequently in the literature
- Cognitive symptoms and fatigue are frequently reported by patients
- Over 50% patients reported cognitive issues and or fatigue in a study using qualitative interviews to elicit patient reported symptoms



CLASSIC SYMPTOMS

Headache /
Head pain
Most commonly orthostatic

Brain Fog / Fatigue other cognitive

Neck pain
Intrascapular pain

Tinnitus other auditory/aural



Dizziness

Lightheaded/disequilibrium

Photophobia other visual

Nausea

REFERENCES

- Schievink WI, Maya MM, Louy C, Moser FG, Tourje J. Diagnostic Criteria for Spontaneous Spinal CSF Leaks and Intracranial Hypotension. American Journal of Neuroradiology 2008;29:853.
- D'Antona L, Jaime Merchan MA, Vassiliou A, et al. Clinical Presentation, Investigation Findings, and Treatment Outcomes of Spontaneous Intracranial Hypotension Syndrome: A Systematic Review and Meta-analysis. JAMA Neurol 2021;78:329-337.
- Amrhein TJ, McFatrich M, Ehle K, et al. Patient experience of spontaneous intracranial hypotension (SIH): qualitative interviews for concept elicitation. J Patient Rep Outcomes 2023;7:82.
- Urbach H, Fung C, Dovi-Akue P, Lützen N, Beck J. Spontaneous Intracranial Hypotension. Dtsch Arztebl Int. 2020; I 17(27-28):480-487.

